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Cardiac Hydatid Cyst of the Interventricular Septum, an Incidental Finding in a 6-Year-Old Girl: Case Report and Review of Literature

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Abstract **Original Research Article**

Cardiac hydatid cysts are rare and represent less than 2% of all hydatid cases. They can occur as part of a widespread systemic infection or as an isolated event. Cardiac hydatic cysts rarely involve the interventricular septum, they also usually evolve slowly, explaining their rarity in children. We report in this work, an incidental finding of a cardiac hydatid cyst located in the interventricular septum in a 6-year-old girl, during an abdominal ultrasound exam performed after minor abdominal trauma.

Keywords: Hydatid cyst, cardiac, children, imaging.

INTRODUCTION

Hydatidosis is a zoonotic parasitic infection caused by tapeworms of the genus Echinococcus. It is a widespread problem in sheep raising countries. Despite current initiatives, Morocco continues to have a high disease burden with substantial socio-economic burden [1].

It commonly involves the liver (50-70%) and lungs (20-30%) [2]. Cardiac involvement is very rare and is usually asymptomatic and slowly evolving explaining its rarity in children [3]. Hydatid cysts of the interventricular septum (IVS) account for less than 4% of all cardiac locations [4].

Here we report a case of incidentally detected cardiac hydatid cyst located in the interventricular septum in a 6-year-old.

CASE REPORT

A 6-year-old female from the rural area of Amizmiz Morocco, with no particular medical history, presented to the emergency room 3 hours after minor abdominal trauma complaining of acute abdominal pain. Physical examination revealed a mild epigastric tenderness. As part of traumatic assessment, an abdominal ultrasound (US) was performed and revealed neither accumulation of free fluid nor any other feature that may be suggestive of organ injury. However, it revealed multiples well defined, thin-walled cystic lesions in the right lobe of the liver with posterior enhancement. One of the lesions demonstrated internal detachment membrane, the so-called water lilly sign. During exam, a large, thin-walled cystic lesion was seen incidentally in the interventricular septum. A transthoracic echocardiography was performed for a better assessement of the cardiac lesion and showed the presence of an anechoic cystic formation at the level of the inflow septum extending all the way up to the mitral annulus and to the coronary sinus posteriorly measuring 52x40mm, this mass also comes in contact with the posterior mitral valve leaflet evoking a HC at first (Figure 1). It also revealed a moderate mitral regurgitation over cleft mitral valve and a non-dilated LV with preserved systolic function. A resection of the mass and a mitral plasty were indicated.

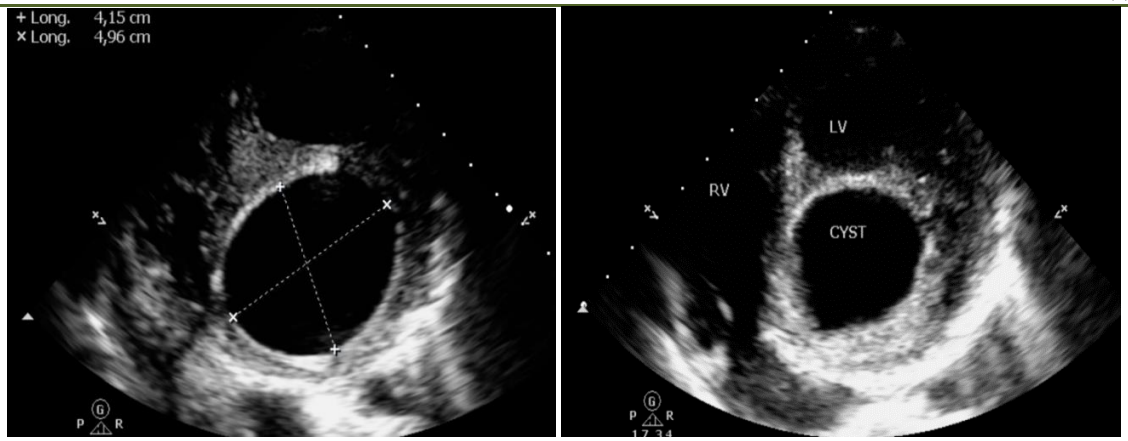

Figure 1: Transthoracic echocardiography images showing an anechoic cystic lesion in the interventricular septum

The patient was then referred for a chest and abdominal CT that revealed similar findings with multiples well defined, thin-walled hypodense cystic lesions, in the right hepatic lobe, non-enhanced after contrast. Some of the lesions demonstrated some peripheral wall calcifications in pre contrast images and another lesion showed internal detachment membrane, compatible with the ultrasound findings. The largest lesion was located in the VI hepatic segment and measured 65 x 57 mm. The cardiac lesion appeared on CT images as a well-defined thin-walled hypodense lesion in the interventricular septum. No solid enhancing components, wall calcification or adjacent infiltrative/inflammatory changes were noted with this lesion (Figure 2).

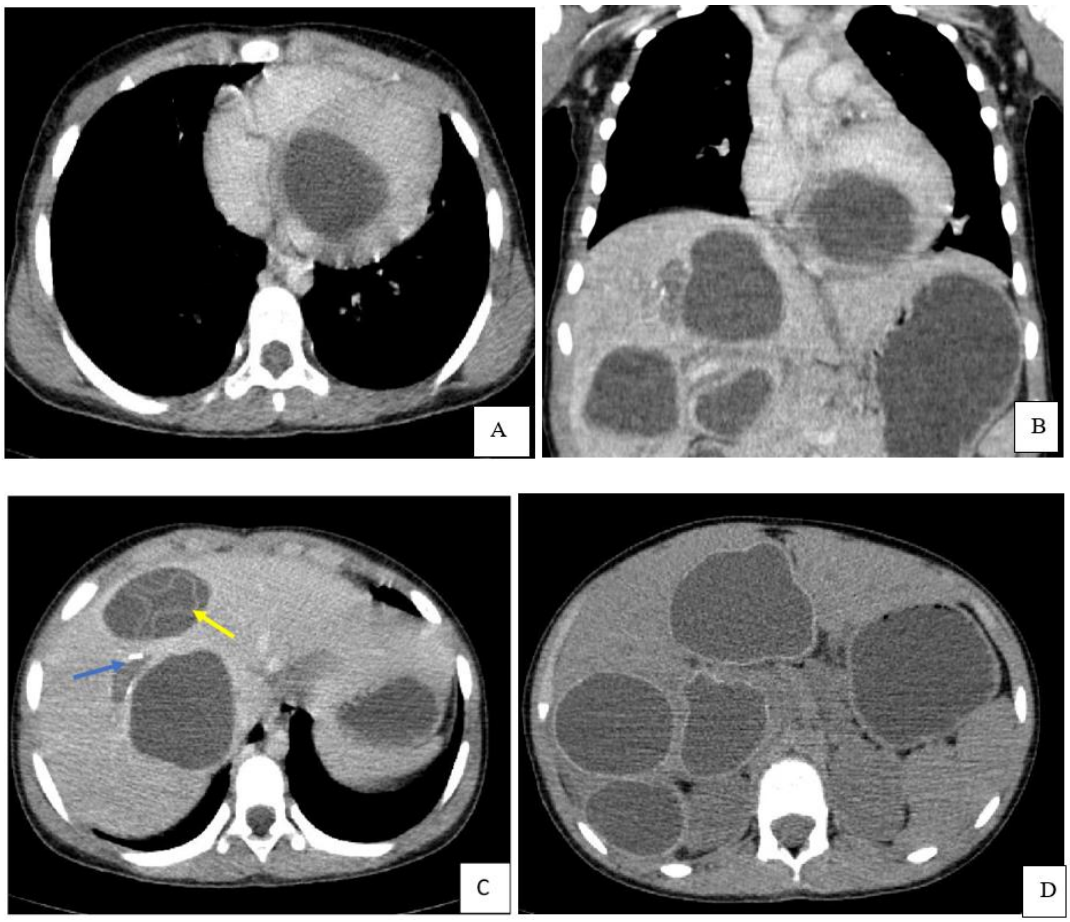

Figure 2: Axial (A, C, D) and coronal (B) thoraco-abdominal CT images with (A, B, C) and without (D) contrast showing multiples well defined, thin-walled cystic hepatic lesions with wall calcifications (blue arrow) and internal detachment membrane (yellow arrow). The cardiac lesion appeared as a well-defined thin-walled cystic lesion in the interventricular septum

Keeping in mind multiplicity of the lesions, typical CT features, contact with animals and geographic location of the patient, the diagnosis of hydatid cysts was made and the patient was prescribed Albendazole 400mg twice daily for 2 months then the cardiac cyst was surgically removed. The patient was seen at 8 weeks follow up and appeared to be doing well and was still taking Albendazole therapy.

Review:

We retrospectively reviewed the medical records of cardiac HCs located in the interventricular septum in patients aged 18 or under, before December 2022 using Pubmed. We used the following searching terms: "Hydatid cyst" and "interventricular septum". We included papers in English, Spanish and French and excluded papers with patients above the age of 18 years old. Twenty-eight patients were included in the review with a mean age of 11.8 years. Eight were females and 20 were males. Most of the cases reviewed were from Turkey (twelve cases) and middle east (5 cases), the others were from Spain (four cases), Kyrgystan (3 cases), India (one case), Tunisia (one case), Ethiopia (one case) and Serbia (one case) (Table 1). Common symptoms were dyspnea, cough, chest pain and fever. Two patients presented with oedema and hepatomegaly and one patient also had lower extremity ischemia. Transthoracic echocardiogram was performed in twenty six patients, 7 patients also received an MRI screening and 4 patients underwent a CT exam. One patient was diagnosed per-operatively and in another patient the diagnosis was established in post mortem after an autopsy. 7 patients had other organ involvement including one patient who had bilateral pulmonary embolism and another who had a femoral artery embolism. The hydatid serology was positive in only 8 patients. Two patients refused surgery and almost all patients underwent a cystectomy under cardio pulmonary bypass. The post- operative use of albendazole was reported in 14 patients and mebendazole was reported in one patient.

At follow-up all patients who underwent surgery were doing well, and only one patient developed a pulmonary hypertension.

Author	De los Arcos *et al*., [5]	Farooki *et al*., [6]	Eren *et al*., [7]	Mandke *et al*., [8]	Unal *et al*., [3]
Year	1971	1976	1989	1991	1995
Age	11	8	18	18	14
Gender	M	M	M	F	F
Country	Spain	Moved from Middle East to USA	Turkey	India	Turkey
Cardiac location	IVS	IVS	IVS	IVS + IA	IVS
Diagnosis	Per-operative	TTE	TTE	Autopsy	TTE
Symptoms	-	-	Palpitations - Shortness of breath	NR	Chest pain
Size (cm)	10	NR	4 x 6	13.6 x 10.2 x 8	3.5 x 4.2
Other organs involvment	NR	Face and neck	NR	No	No
Serology	NR	NR	NR	-	Positive
Medication	NR	NR	NR	-	Mebendazole
Surgery	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Autopsy	Refused surgery
Histology	NR	Yes	NR	Yes	No
Follow up	NR	Uneventful	Uneventful	Death	NR

Ben Hamda *et al.*, [14]	Yalçin *et al.*, [3]	S *et al.*, [13]	S *et al.*, [13]	S *et al.*, [13]	Ozer *et al.*, [12]	Onursal *et al.*, [11]	Perez Martinez *et al.*, [10]	Maroto *et al.*, [9]	Lopez Rioz *et al.*, [3]
2002	2002	2001	2001	2001	2001	2001	1999	1998	1997
9	11	9	10	12	17	13	3	3.5	4
M	M	F	M	F	M	M	M	M	M
Tunisia	Turkey	Kyrgyzstan	Kyrgyzstan	Kyrgyzstan	Turkey	Turkey	Spain	Spain	Spain
IVS + RV	IVS	IVS	IVS + RV	IVS + RV	IVS	IVS	IVS	IVS	IVS
TTE	TTE	TTE	TTE	TTE	TTE	TTE	TTE - MRI	TTE	TTE
-	Dyspnea- Cough-Haemoptysis-Fever	Dyspnea-Weakness	Oedema-Hepatomegaly - Weakness	Oedema-Hepatomegaly	Cough-Dyspnea	Fever- Cough-Dyspnea-Chest pain	-	-	-
4 X 4	2	5	3.5	3	NR	NR	3.4 x 2.8	3 x 2	3.4 x 2.8
Liver and lung	Multiorgan involvment	No	No	No	No	No	No	No	No
Positive	Positive	NR	NR	NR	NR	NR	Negative	Negative	Negative
NR	Albendazole	Albendazole	Albendazole	Albendazole	NR	Albendazole	NR	NR	NR
Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB
Yes	NR	Yes	Yes	Yes	NR	NR	Yes	Yes	NR
Uneventful	Uneventful	Uneventful	Uneventful	Uneventful	Pulmonary hypertension	Uneventful	Uneventful	Uneventful	Uneventful

Tefera *et al.*, [22]	Sahin *et al.*, [21]	Goçen *et al.*, [20]	Mirzae *et al.*, [19]	Tuncer *et al.*, [18]	Mohsen *et al.*, [17]	Mohsen *et al.*, [17]	Ilic *et al.*, [16]	Ugurlucan *et al.*, [15]
2016	2015	2014	2014	2010	2008	2008	2008	2006
9	17	6	14	17	4	5	13	12
F	M	F	M	M	M	M	M	F
Ethiopia	Turkey	Turkey	Iran	Turkey	Egypt	Egypt	Serbia	Turkey
IVS	IVS	IVS	IVS	IVS	IVS	IVS	IVS	IVS + Pericardium
TTE	TTE - TTO - CT	TTE	TTE	TTE	TTE - MRI	TTE - MRI	TTE	TTE - MRI
Palpitations - Weakness	Palpitations - Dizziness	Chest pain-Dyspnea	Acute low extremity pain	Chest pain	Palpiations-Dyspnea	Dyspnea-Weakness	Chest pain-Dyspnea- Rash-Fever	Acute dyspnea-palpitations
3.8 x 3.4	6.6 x 3.5	4 x 3	4 x 3.5	5 x 5	3 x 3	2.1 x 2.3	3.3 x 2.1	5.7 x 5.3 x 4.5
No	No	No	Femoral artery	No	Liver	No	Bilateral pulmonary	No
NR	Positive	NR	Positive	NR	NR	NR	Positive	NR
Albendazole	Albendazole	Albendazole	Albendazole	NR	Albendazole	Albendazole	Albendazole	NR
Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB	Cystectomy CPB-Embolectomy	Cystectomy CPB
NR	Yes	NR	Yes	NR	NR	NR	Yes	Yes
Uneventful	Uneventful	Uneventful	Uneventful	Uneventful	Uneventful	Uneventful	Uneventful	NR

Ibrahim *et al*., [26]	2020	15	M	Iraq	IVS	TTE - CT	Dyspnea- Weakness- Right upper abdominal pain	7.5 x 5.5	Liver	Positive	NR	Cystectomy CPB	Yes	Uneventful
Iriz *et al*., [25]	2020	15	F	Turkey	IVS	TTE - CT -MRI	Chest pain	5.7 x 4.4 x 4.2	No	Negative	Albendazole	Cystectomy CPB	Yes	Uneventful
Iyigun *et al*., [24]	2020	18	M	Turkey	IVS	TTE - TTO - MRI	-	7.4 x 4.7 x 4.7	No	NR	Albendazole	Cystectomy CPB	Yes	Uneventful
Alis and Turna [23]	2018	18	M	Turkey	IVS	TTE - CT -MRI	Chest pain- Dyspnea	6.5 x 5.5 x 5.5	No	Positive	Refused treatment	Refused surgery	No	No

DISCUSSION

Hydatid disease is a parasitic infection caused by larvae of Echinococcus granulosus, which is still endemic in Morocco and many other sheep-raising countries. Dogs and other carnivores are the primary host and sheep the intermediate host, whereas humans are the accidental intermediate host. Humans usually become affected by the ingestion of food or water contaminated by embryonated eggs shed in dogs' faeces [27, 28].

Cardiac localization of hydatid cyst is very rare and represents 0.5 – 2% of all patients with hydatidosis. Cardiac involvement occurs from the systemic or pulmonary circulation or by direct extension from adjacent structures [29]. The left ventricle is the most frequent site of infection accounting for 55 to 60 % of cardiac hydatid cysts, followed by the right ventricle (10-15%), pericardium (7%), pulmonary artery (6%–7%), left atrium (6%–8%), right atrium (3%–4%), and interventricular septum (4%). Cardiac echinococcosis usually stays asymptomatic for years and only about 10% of patients present symptoms in the early stages [30]. The clinical presentation varies depending on the site and the size of the cysts. Pre-cordial pain is the most common symptom. Valvular dysfunction and pulmonary hypertension have also been reported [4]. Complications such as cardiac tamponade by rupture in the pericardial sac, anaphylactic shock and embolisation into pulmonary and systemic sites can occur. Early diagnosis of this condition is crucial to avoid these complications. Occasionally, these cysts have been reported to cause coronary artery compression [28].

The chest radiograph can often appear normal, as seen in our case. Depending upon the location, size and stage of the cyst, abnormalities of the cardiac contour and calcifications may be noted. Echocardiography is a sensitive and specific tool to diagnose hydatid cysts in terms of location, size, and to calculate the pressure on the vital parts of the heart [31]. The non-invasive nature of the modality and lack of cardiac motion artefacts (which both occur with CT and MRI) make echocardiography the modality of choice in imaging cardiac hydatid disease [28]. Computed tomography and MRI provide further information, such as the extent and anatomic relationships of the cysts. CT also best shows wall calcifications and MRI depicts the exact anatomic location and the nature of the internal and external structures. The presence of a multivesicular cystic lesion or membrane detachment is highly indicative of hydatidosis rather than the multiple differential diagnosis of cardiac cystic lesions [4]. Serologic tests can be false-negative in 10% to 20% of patients with hepatic hydatid cysts, 40% with pulmonary cysts, and 50% with cardiac cysts; this is most likely linked with an insufficient immune response and as leakage of cyst fluid is believed

to be essential for immunosensitization, an unruptured cyst can result in a false-negative result [30].

Surgical excision on cardiopulmonary bypass is the treatment of choice for cardiac hydatid disease even in asymptomatic patients aided by Albendazole therapy (400 mg twice daily) that is typically prescribed for at least 4 days preoperatively and for 4 to 12 weeks postoperatively [30, 32]. Conservative therapy has been considered in a few cases on high risk patients or those with completely calcified cysts and no haemodynamic complications [4, 33].

CONCLUSION

In conclusion, hydatid cyst of the heart and specifically the interventricular septum is rare. Awarness of this condition is crucial for both clinicians and radiologists. Echocardiography, CT, and MRI are useful in the diagnosis and location of cardiac echinococcosis. Treatment consisting of surgery with concurrent albendazole therapy typically yields excellent results. Early diagnosis of this condition is essential to avoid fatal complications.

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